Gastrointestinal stromal tumour (GIST) arising in a colonic duplication cyst: case report

doi:10.1111/j.1463-1318.2009.02095.x

Case report

A 31-year-old woman presented with acute diffuse abdominal pain. Computerized tomography (CT) revealed an oedematous, slightly irregular 5.8 × 3.6 × 5.6 cm mass in the pelvis, behind the uterus, with a hypercaptating wall and inlying hypocaptating nodules. A differential diagnosis of pelvic inflammatory disease and ovarian torsion was considered.

At laparoscopy, an inflamed mass, adherent to the jejunum and sigmoid, was seen in the right iliac fossa. The mass was removed by sigmoid resection.

On gross examination, the lesion was 6 cm in diameter and showed a central cavity filled with blood. There was no communication between the lumen of the adherent sigmoid and the cavity in the mass.

Microscopically (Fig. 1), the mass consisted of uniform, densely packed, diffuse sheets of spindle cells with little atypia and a mitotic count of < 5/50 high-power fields. The cells stained positively with antibodies directed against c-kit (CD 117), CD 34 and alpha-smooth muscle actin (alpha-SMA), but S-100 and desmin were negative. The diagnosis of a gastrointestinal stromal tumour (GIST) was made. Following the National Institute of Health Risk Stratification Categories [1], the GIST had an intermediate malignant potential: the tumour was 6 cm (intermediate) with a mitotic count of < 5/50 HPF (low). The inner surface of the macroscopically observed central cavity was lined by a colon-like mucosa with underlying remnants of smooth muscle. The final histopathological diagnosis was a GIST arising in a duplication cyst of the colon.

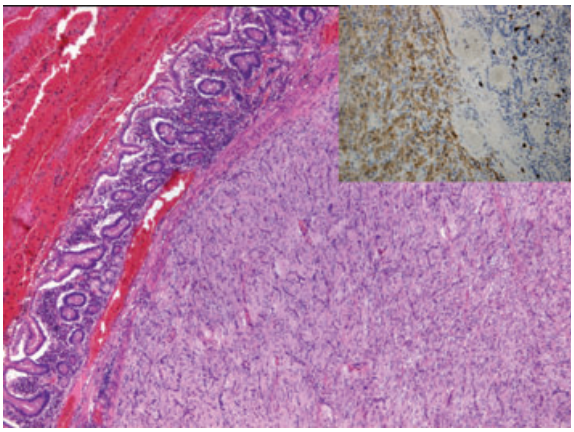

Figure 1 Central cavity lined by a colon-like mucosa with underlying remnants of smooth muscle and tumoral mass consisted of uniform, densely packed, diffuse sheets of spindle cells with little atypia (HE × 50). Immunohistochemistry (inset) shows the spindle cells to express c-kit (CD 117).

Discussion

Alimentary tract duplications are uncommon developmental anomalies. They were first described in 1884 as spherical or tubular structures firmly attached to, and sometimes in communication with, a part of the gastrointestinal tract [2]. They can occur anywhere along the gastrointestinal tract. The colon is with 6–8% of all duplications the least common site [3].

There are several reports of malignant degeneration of duplication cysts in adult patients. In most cases, it concerned adenocarcinoma, although a few cases of squamous cell carcinoma have also been described [4]. A GIST arising in a colonic duplication cyst has never been reported in the English literature.

Gastrointestinal stromal tumours are the most common mesenchymal tumours of the gastrointestinal tract, and they are believed to arise from the precursor cell of the interstitial cell of Cajal. Less than 5% of GISTs is present in colorectum [5]. Rare cases have also been reported in the mesentery, omentum and retroperitoneum. Standard treatment of GIST is surgical resection. Imatinib mesylate is the standard regimen if the tumour is metastatic or unresectable.

The present case illustrates that the rare diagnosis of an enteric duplication cyst should be considered in the differential diagnosis of abdominal mass. Because of the uncommon but well-documented risk of malignant degeneration, resection is indicated when a duplication cyst is suspected in an adult. In our patient, a complete surgical resection was performed and no adjuvant treatment was started.

A. S. Van Rompuy*, M. Lannoo†, M. D'hondt†, A.D'Hoore† and K. Geboes*

Department of *Pathology and †Abdominal Surgery, University Hospitals KU Leuven, Blegium

Correspondence to: Karel Geboes, Department of Pathology, University Hospitals KU Leuven, Campus St. Rafaël, Minderbroedersstraat 12, Belgium.
E-mail: karel.geboes@uz.kuleuven.ac.be

Received 9 July 2009; accepted 4 August 2009; Accepted Article online 30 October 2009

References

1 Fletcher CD, Berman JJ, Corless C, *et al.* Diagnosis of gastrointestinal stromal tumours: a consensus approach. *Hum Pathol* 2002; **33:** 459–65.
2 Fitz RH. Persistent omphalo-mesenteric remains: their importance in the causation of intestinal duplication, cyst formation and obstruction. *Am J Med Sci* 1884; **80:** 30–57.
3 Anderson MC, Silberman WW, Shields TW. Duplication of the alimentary tract in the adult. *Arch Surg* 1962; **85:** 94–124.
4 Inoue Y, Nakamura H. Adenocarcinoma arising in colonic duplication cysts with calcification: CT findings of two cases. *Abdom Imaging* 1998; **23:** 135–7.
5 Miettinen M, Lasota J. Gastrointestinal stromal tumors (GISTs): definition, occurrence pathology, differential diagnosis and molecular genetics. *Pol J Pathol* 2003; **54:** 3–24.